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OLGU SUNUMU

Giant hemangiopericytoma of the neck: a case report

Dev boyun hemanjiyoperistomu: Olgu sunumu

Yahya GÜLDİKEN,[1] Emre A. YÜCEL,[1] Mustafa ÖZDEMİR,[1] Bilge BİLGİÇ[2]

Hemangiopericytomas are rare vascular tumors and one-thirds occur in the head and neck. These tumors grow slowly and may easily be mistaken for a benign lesion. A twenty-nine-year-old man presented with a large mass on the left side of his neck. Palpation, ultrasonography, and magnetic resonance imaging findings were incorporated into a histopathologic diagnosis of malignant hemangiopericytoma. The tumor was subtotally excised following preoperative embolization of the branches of the subclavian artery and the left vertebral artery. The patient was submitted to radiotherapy. He has been under close follow-up for 16 months without any recurrences.

Key Words: Head and neck neoplasms/pathology; hemangiopericytoma/pathology/diagnosis/surgery.

Hemanjiyoperistomlar nadir vasküler tümörlerdir; 1/3'ü baş-boyun bölgesinde görülür. Yavaş büyüme gösteren bu tümörler kolaylıkla benign lezyonlarla karıştırılabilir. Boyun sol tarafında geniş kitle ile başvuran 29 yaşındaki bir erkek hastada, palpasyon, ultrasonografi ve manyetik rezonans görüntülemesini takiben histopatolojik inceleme ile malign hemanjiyoperistom tanısı kondu. Subklavyen ve sol vertebral arter dallarının ameliyat öncesinde embolizasyonu yapıldı. Tümörün subtotal eksizyonunun ardından hasta radyoterapiye sevk edildi. On altı aydır izlenen olguda rekürense ya da herhangi bir komplikasyona rastlanmadı.

Anahtar Sözcükler: Baş ve boyun neoplazmları; hemangioperisitoma/patoloji/tanı/cerrahi.

Hemangiopericytomas (HPC) constitute an unusual group of mesenchymal vascular neoplasms, originating from capillary pericytes of Zimmerman. They may be detected at any site where capillaries are found. Frequently, they arise from musculoskeletal tissues and the skin, but they may also originate from viscera.[1] Generally, they grow slowly and are locally infiltrative with a varying malignant potential.[2] They account for more than 1% of all vascular neoplasms and approximately one-thirds is encountered in the head and neck.[1]

We hereby report a case of giant and locally aggressive HPC in the neck.

CASE REPORT

A twenty-nine-year-old man was referred to our department, with a large mass on the left side of his neck. The mass was first noted by the patient six months ago. He was examined elsewhere and followed-up with a diagnosis of a benign lesion. However, the lesion underwent a steady growth reaching a size of 15x7 cm in diameter, with rubbery

◆ *Departments of [1]Otolaryngology and [2]Pathology, Medicine Faculty of İstanbul University, İstanbul - Turkey.*

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◆ *Correspondence: Dr. Emre A. Yücel. Millet Cad. Özbek Süleyman Sok., Aras Apt., No: 8 D: 27, 34270 Fındıkzade, İstanbul, Turkey.*
Tel: +90 212 - 585 71 32 Fax: +90 212 - 534 71 33
e-mail: eayucel@superonline.com

◆ *İstanbul Üniversitesi İstanbul Tıp Fakültesi, [1]Kulak Burun Boğaz Hastalıkları Anabilim Dalı, [2]Patoloji Anabilim Dalı, İstanbul.*

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◆ *İletişim adresi: Dr. Emre A. Yücel. Millet Cad. Özbek Süleyman Sok. Aras Apt., No: 8 D: 27, 34270 Fındıkzade, İstanbul.*
Tel: 0212 - 585 71 32 Faks: 0212 - 534 71 33
e-posta: eayucel@superonline.com

consistency and fixed, sharp contours. On palpation, lateral and posterior displacements of the carotid artery and the sternomastoid muscle were elicited, respectively. The carotid artery was felt subcutaneously. He was complaining of numbness in his left arm, although his muscle strength was normal. The development of an ipsilateral Horner's syndrome was noted. Ultrasonography revealed a solid mass in relation to the left thyroid lobe. Fine-needle aspiration biopsy failed to allow a diagnosis. Magnetic resonance imaging showed a mass lesion between the left submandibular area and the superior mediastinum, measuring 13.5x7x6 cm. Necrotic areas and vascular spaces were identified in the center of the mass, showing nonhomogeneous contrast enhancement. T_1-weighted images demonstrated hypointense signals, whereas T_2-weighted images showed high signal intensities. An incisional biopsy was performed under general anesthesia. The anterior surface of the lesion was encapsulated and profuse bleeding was noted. Histopathologic examination of the specimen revealed malignant HPC.

Considering the aggressive nature of the tumor, radical neck dissection was planned. After obtaining informed consent from the patient, preoperative embolization of the major feeding vessels were performed, including the branches of the subclavian artery and the left vertebral artery. Perioperatively, no encapsulation was seen on the posterior surface of the tumor. The jugular vein was obliterated. The vagal and accessory nerves were sacrificed because of tumor invasion. Invasion to the deep neck muscles, prevertebral fascia, and even to the intervertebral spaces was also noted. The tumor was subtotally removed without marked bleeding.

Fig. 1 - *A large bulky lesion on the left side of the neck.*

Macroscopically, the mass measured 4x7x18 cm. The pseudocapsule surrounding the tumor was not intact. On cut section the tumor appeared grey-white, showing hemorrhagic areas. Histopathologic examination showed infiltration of oval-spindle cells with indistinct cytoplasm. The vasculature was prominent, consisting of capillary vessels. There were solid, cellular areas. Mitotic activity was 4-5/10 high power field. Pleomorphism, atypical mitotic appearance, areas of necrosis, and hemorrhage were

(a)

Fig. 2 - ***(a)*** *Axial and* ***(b)*** *sagittal magnetic resonance views showing necrotic areas and involvement of the intervertebral space (arrow).*

observed. Immunohistochemically, the tumoral cells stained positively for factor XIIIa. The endothelial cells stained positively for CD31. With the diagnosis of malignant HPC, the patient was referred to the radiotherapy department for full-course of radiation. He has been under close follow-up for 16 months without any recurrences.

DISCUSSION

Hemangiopericytoma is an uncommon mesenchymal vascular neoplasm accounting for only 1% of adult vascular head and neck tumors.[2] It may be defined as sarcomas, and as with other sarcomas, survival is correlated with the grade, size, and the margin status.[3] Enzinger and Smith[4] analyzed 106 cases of HPC and found that 16% was located in the head and neck region with an overall survival of 70%. Their criterion for malignancy was defined as the appearance of four or more mitotic figures per high power field. They also reported that 10-year-survival decreased from 92% to 63% in patients presenting with tumors greater than 6.5 cm in diameter.[4]

Hemangiopericytoma typically presents as a slowly-growing solitary, asymptomatic mass. There is usually no pain unless local invasion to the deep tissues occurs. Therefore, it may clinically be mistaken for a lipoma and may not be removed, progressing to the final appearance of a giant tumor.[5] This was the case in our patient; he received treatment at another clinic for sometime, during which the mass changed into a bulky tumor, making it impossible to be removed with clear margins.

The most common localizations of HPC are the lower extremities, pelvis, trunk, retroperitoneum, and the head and neck. According to Walike and Bailey,[1] one-thirds of all hemangiopericytomas occurs in the head and neck. They have been reported in the middle ear,[6,7] tongue,[8] nasopharynx,[9] infratemporal fossa,[10] parotid gland,[11] larynx,[12] maxillary sinus,[3] and the neck.[13,14] There is no sex and age predominance; however, a slight peak has been reported for the middle ages.[11] The clinical course of HPC is unpredictable.[13] Of these locally aggressive tumors, 50% has a predilection for recurrence, and 20 to 45% for distant metastasis. Walike and Bailey,[1] reported 40% recurrence and 10% distant metastasis in 45 patients with head and neck HPC.[1] Seibert[9] reported a total recurrence rate of 50%, and a 10-year survival rate of 70%. Sixty-nine percent of recurrences are associated with metastasis.[15] The mode of metastasis is primarily vascular, and the lungs are the most common site.

The choice of treatment is wide surgical resection with adequate margins.[2] The absence of encapsulation represents a major problem for the surgeon. Taking high vascularity into consideration, preoperative embolization may be performed.[10] High-grade lesions and inadequate surgical margins may necessitate postoperative radiotherapy,[3] although some authors drew attention to the tendency of HPC to be

Fig. 3 - *Solid area of pleomorphic oval-spindle cells with indistinct cytoplasm, scattered small capillary vessels (H-E x 500).*

resistant to radiotherapy.[12] Moreover, chemotherapy may be considered in metastatic disease, because favorable results have been reported with cyclophosphamide, vincristine, dacarbazine, methotrexate, or dactinomycin.[10] Recent use of interferon-alpha has been associated with stabilization of the lesions in patients with metastatic disease.[3]

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